



8p11 myeloproliferative syndrome

8p11 myeloproliferative syndrome is a blood cancer that involves different types of blood cells. Blood cells are divided into several groups (lineages) based on the type of early cell from which they are descended. Two of these lineages are myeloid cells and lymphoid cells. Individuals with 8p11 myeloproliferative syndrome can develop both myeloid cell cancer and lymphoid cell cancer.

The condition can occur at any age. It usually begins as a myeloproliferative disorder, which is characterized by a high number of white blood cells (leukocytes). Most affected individuals also have an excess of myeloid cells known as eosinophils (eosinophilia).

In addition to a myeloproliferative disorder, many people with 8p11 myeloproliferative syndrome develop lymphoma, which is a form of blood cancer that involves lymphoid cells. The cancerous lymphoid cells grow and divide in lymph nodes, forming a tumor that enlarges the lymph nodes. In most cases of 8p11 myeloproliferative syndrome, the cancerous cells are lymphoid cells called T cells. Lymphoma can develop at the same time as the myeloproliferative disorder or later.

In most people with 8p11 myeloproliferative syndrome, the myeloproliferative disorder develops into a fast-growing blood cancer called acute myeloid leukemia.

The rapid myeloid and lymphoid cell production caused by these cancers results in enlargement of the spleen and liver (splenomegaly and hepatomegaly, respectively). Most people with 8p11 myeloproliferative syndrome have symptoms such as fatigue or night sweats. Some affected individuals have no symptoms, and the condition is discovered through routine blood tests.

Frequency

The prevalence of 8p11 myeloproliferative syndrome is unknown. It is thought to be a rare condition.

Genetic Changes

8p11 myeloproliferative syndrome is caused by rearrangements of genetic material (translocations) between two chromosomes. All of the translocations that cause this condition involve the *FGFR1* gene, which is found on the short (p) arm of chromosome 8 at a position described as p11. The translocations lead to fusion of part of the *FGFR1* gene with part of another gene; the most common partner gene is *ZMYM2* on chromosome 13. These genetic changes are found only in cancer cells.

The protein normally produced from the *FGFR1* gene can trigger a cascade of chemical reactions that instruct the cell to undergo certain changes, such as growing

and dividing. This signaling is turned on when the FGFR1 protein interacts with growth factors. In contrast, when the *FGFR1* gene is fused with another gene, FGFR1 signaling is turned on without the need for stimulation by growth factors. The uncontrolled signaling promotes continuous cell growth and division, leading to cancer.

Researchers believe the mutations that cause this condition occur in a very early blood cell called a stem cell that has the ability to mature into either a myeloid cell or a lymphoid cell. For this reason, this condition is sometimes referred to as stem cell leukemia/lymphoma.

Inheritance Pattern

This condition is generally not inherited but arises from a mutation in the body's cells that occurs after conception. This alteration is called a somatic mutation.

Other Names for This Condition

- 8p11 stem cell leukemia/lymphoma syndrome
- 8p11 stem cell syndrome
- myeloid and lymphoid neoplasms with FGFR1 abnormalities
- stem cell leukemia/lymphoma

Diagnosis & Management

Genetic Testing

- Genetic Testing Registry: Chromosome 8p11 myeloproliferative syndrome
<https://www.ncbi.nlm.nih.gov/gtr/conditions/C3150773/>

Other Diagnosis and Management Resources

- Cancer.Net from the American Society of Clinical Oncology: Acute Myeloid Leukemia Diagnosis
<http://www.cancer.net/cancer-types/leukemia-acute-myeloid-aml/diagnosis>
- Cancer.Net from the American Society of Clinical Oncology: Acute Myeloid Leukemia Treatment Options
<http://www.cancer.net/cancer-types/leukemia-acute-myeloid-aml/treatment-options>
- Cancer.Net from the American Society of Clinical Oncology: Non-Hodgkin Lymphoma Diagnosis
<http://www.cancer.net/cancer-types/lymphoma-non-hodgkin/diagnosis>
- Cancer.Net from the American Society of Clinical Oncology: Non-Hodgkin Lymphoma Treatment Options
<http://www.cancer.net/cancer-types/lymphoma-non-hodgkin/treatment-options>

General Information from MedlinePlus

- Diagnostic Tests
<https://medlineplus.gov/diagnostictests.html>
- Drug Therapy
<https://medlineplus.gov/drugtherapy.html>
- Genetic Counseling
<https://medlineplus.gov/geneticcounseling.html>
- Palliative Care
<https://medlineplus.gov/palliativecare.html>
- Surgery and Rehabilitation
<https://medlineplus.gov/surgeryandrehabilitation.html>

Additional Information & Resources

MedlinePlus

- Health Topic: Acute Myeloid Leukemia
<https://medlineplus.gov/acute-myeloid-leukemia.html>
- Health Topic: Blood Disorders
<https://medlineplus.gov/blood-disorders.html>
- Health Topic: Bone Marrow Diseases
<https://medlineplus.gov/bone-marrow-diseases.html>
- Health Topic: Lymphoma
<https://medlineplus.gov/lymphoma.html>

Additional NIH Resources

- National Cancer Institute: Adult Acute Myeloid Leukemia
<https://www.cancer.gov/types/leukemia/patient/adult-aml-treatment-pdq>

Educational Resources

- Atlas of Genetics and Cytogenetics in Oncology and Haematology
<http://atlasgeneticsoncology.org/Anomalies/8p11inMPDID1091.html>
- Cedars-Sinai: Leukemia
<http://www.cedars-sinai.edu/Patients/Health-Conditions/Leukemia.aspx>
- Johns Hopkins Medicine: Non-Hodgkin's Lymphoma
http://www.hopkinsmedicine.org/kimmel_cancer_center/types_cancer/non_hodgkin_lymphoma.html
- KidsHealth from Nemours: Leukemia
<http://kidshealth.org/en/parents/cancer-leukemia.html>

- MalaCards: 8p11 myeloproliferative syndrome
http://www.malacards.org/card/8p11_myeloproliferative_syndrome
- Orphanet: Myeloid neoplasm associated with FGFR1 rearrangement
http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=168953

Patient Support and Advocacy Resources

- American Cancer Society
<https://www.cancer.org/>
- Leukemia and Lymphoma Society
<http://www.lls.org/>

Scientific Articles on PubMed

- PubMed
<https://www.ncbi.nlm.nih.gov/pubmed?term=%28Myeloproliferative+Disorders%5BMAJR%5D%29+AND+%28%288p11+myeloproliferative+syndrome%5BTIAB%5D%29+OR+%28stem+cell+leukemia/lymphoma%5BTIAB%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+3600+days%22%5Bdp%5D>

OMIM

- CHROMOSOME 8p11 MYELOPROLIFERATIVE SYNDROME
<http://omim.org/entry/613523>

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